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REVERSIBLE POSTERIOR ENCEPHALOPATHY SYNDROME: A LITERATURE REVIEW

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Abstract: Posterior reversible encephalopathy syndrome (PRES) is a syndrome with clinical and radiological manifestations. Although it occurs more frequently in the context of hypertensive crisis, pre-eclampsia or the use of immunosuppressive medications, PRES manifests itself in many clinical contexts. Typical signs and symptoms include headache, mental confusion, visual symptoms, and seizures, associated with findings consistent with vasogenic edema in the subcortical white matter with a predominance in the posterior cerebral hemispheres seen on neuroimaging exams. Treatment of arterial hypertension is the mainstay of treatment and recovery tends to occur within two weeks, but a small proportion of patients remain with residual neurological deficits. The immediate identification and early treatment of PRES are essential in preventing permanent damage.
Keywords: Posterior Reversible Encephalopathy Syndrome, Radiology, Neurology, Rheumatology.

INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) is a syndrome with clinical and radiological manifestations, presenting different etiologies grouped together due to similar findings in neuroimaging exams (ANDERSON RC, et al., 2020; HINDUJA A, 2020; CUI HW, et al. al., 2019). It is also commonly called posterior reversible cerebral edema or posterior leukoencephalopathy syndrome. Despite the nomenclature, it is necessary to emphasize that PRES syndrome is not always reversible or restricts the involvement only to the posterior brain regions (HINDUJA A, 2020; FISCHER M & SCHMUTZHARD E, 2017).

PRES was described in 1996 as a syndrome consisting of headache, mental confusion or reduced level of consciousness, visual changes, seizures, and neuroimaging findings characteristic of posterior cerebral white matter edema (CUI HW, et al., 2019; RACCHIUSA S, et al., 2019; FITTRO K & DIZON R, 2018). It is most commonly associated with hypertensive encephalopathy, eclampsia and the use of cytotoxic and immunosuppressive medications, but it has been described in association with several medical conditions (SAAD AF, et al., 2019; MCDERMOTT M, et al., 2018; FUGATE JE & RABINSTEIN AA, 2015).

The immediate identification and early treatment of PRES are essential in preventing permanent damage (GAO B, et al., 2018; LEGRIEL S, et al., 2011). Given this context, this article aimed to carry out a review of the international literature related to PRES, seeking to identify the clinical profile of patients and treatment, prevention and health promotion strategies.

METHODOLOGY

This integrative review was developed in three stages, namely (I) planning, in which the research guidelines were defined, (II) conducting, which consisted of performing the search and selection of articles and files according to the criteria of inclusion and exclusion, and (III) synthesis and analysis of data.

The articles and documents used in this bibliographic review were searched in the National Library of Medicine (PubMED), Academic Google and Latin American and Caribbean Health Sciences (LILACS) databases and published between 2000 and 2021. The words The keywords used in the search were "Posterior Reversible Encephalopathy Syndrome", "Radiology", "Neurology" and "Rheumatology".

Regarding the inclusion criteria, scientific articles, bulletins and protocols from medical societies that presented data on PRES and its repercussions in the scope of human health were selected.

As for the exclusion criteria, duplicated works were disregarded and that, despite having the chosen keywords, did not address the research theme directly, as verified by reading the abstracts. The material search stage resulted in the finding of 103 files. After applying the inclusion and exclusion criteria, 16 works were selected.

LITERATURE REVIEW

Several clinical conditions have been associated as possible etiologies of PRES, with emphasis on autoimmune diseases, which may be related to PRES both because of its side effects on blood pressure and due to the use of immunosuppressive medications during treatment (HINDUJA A, 2020; BARTYNSKI; WS, 2008; LIMAN TG, et al., 2019).

Regarding systemic arterial hypertension, it is believed to be present in most patients with PRES (FITTRO K & DIZON R, 2018; BARTYNSKI WS, 2008; MILLER R, et al., 2021). Despite this, some studies report that up to half of patients do not have severe hypertension and emphasize that an acute rise in blood pressure may be more relevant to the development of the syndrome than chronic arterial hypertension (FUGATE JE & RABINSTEIN AA, 2015; ROTH C & FERBERT A, 2011). Furthermore, it is known that PRES can also occur in normotensive patients in about a quarter of cases (ISCHER M & SCHMUTZHARD E, 2017; STAYKOV D & SCHWAB S, 2012).

Other comorbidities commonly related to PRES include systemic lupus erythematosus, cryoglobulinemia, thrombotic thrombocytopenic purpura, hemolytic-uremic syndrome, in addition to the use of medications such as cyclosporine or cisplatin (HINDUJA A, 2020; MCDERMOTT M, et al., 2018; LIMAN TG, et al., et al., 2018; LIMAN TG, et al., et al., 2018; al., 2019). The signs and symptoms of PRES manifest rapidly over hours to days and the hypertensive crisis may precede the neurological syndrome (SAAD AF, et al., 2019; MCDERMOTT M, et al., 2018; LEGRIEL S, et al, 2011).

Headache tends to be constant, not localized, moderate to severe in intensity and with low response to analgesia. Changes in consciousness, on the other hand, range from mild somnolence to agitation, which may progress to coma in a small proportion of individuals. Visual changes are frequently reported, including hemianopsia, auras and hallucinations. Seizures are mainly tonic-clonic and occur in most patients (ANDERSON RC, et al., 2020; FUGATE JE & RABINSTEIN AA, 2015).

Neuroimaging tests, such as computed tomography (CT) and magnetic resonance imaging (MRI), are essential for the diagnosis of this syndrome. As neuroradiographic alterations are not specific to PRES, a repetition of the exam may be necessary. After instituting treatment, resolution of imaging findings is expected in days to weeks (SAAD AF, et al., 2019; RACCHIUSA S, et al., 2019; GAO B, et al., 2018).

The most characteristic findings on imaging studies include bilateral areas of white matter edema in the posterior cerebral hemispheres. Frontal lobe lesions can be identified in some cases, but they usually also have associated edema in the posterior cerebral hemispheres (Figure 1). Edemas in other regions, such as the superior frontal gyrus, temporal lobe, basal ganglia, cerebellum and brainstem, have already been described in a smaller number of patients (HINDUJA A, 2020; FITTRO K & DIZON R, 2018; MILLER R, et al. al., 2021).

Treatment guidelines are based on the associated clinical conditions in each case. In the face of convulsive conditions, anticonvulsants must be promptly administered and a reduction in the dose of cytotoxic or immunosuppressive

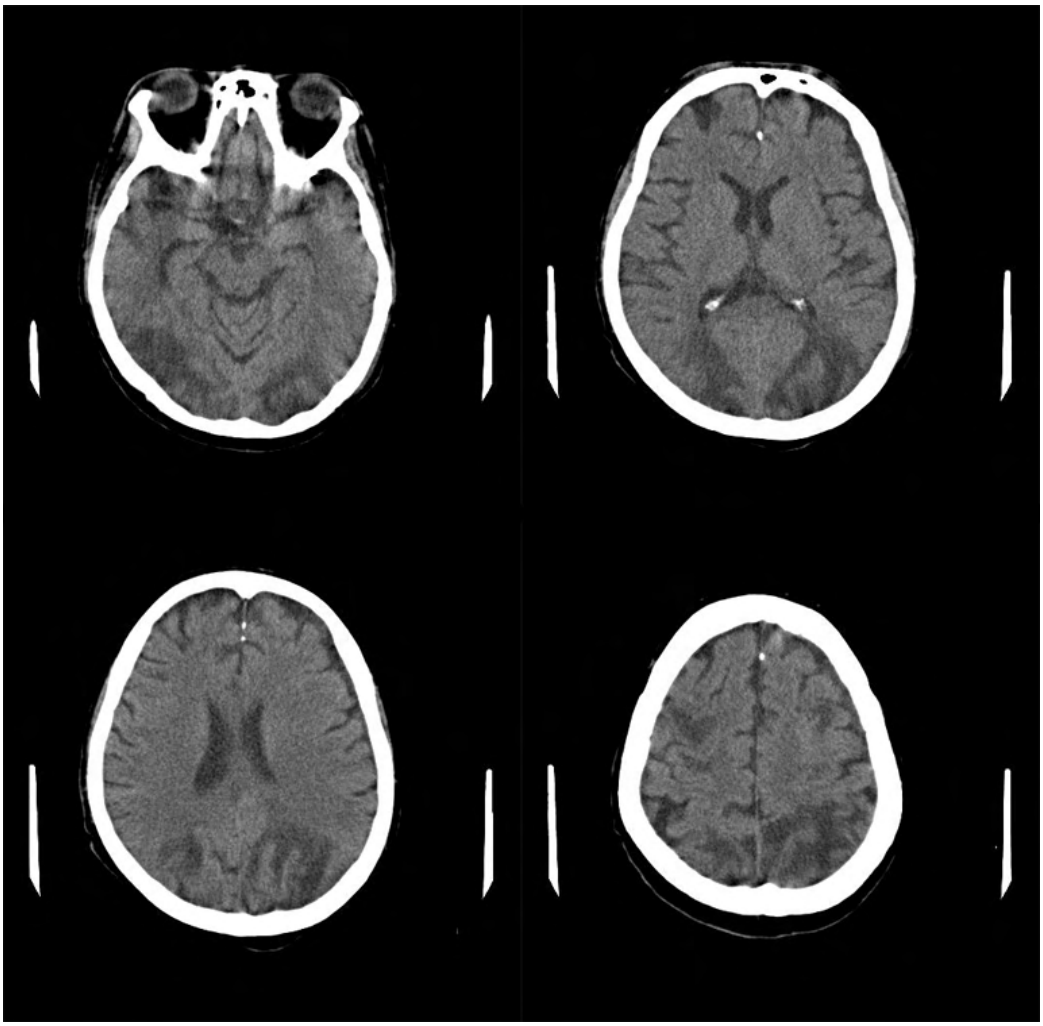

Figure 1 - Axial cranial CT sections showing hypodensity compatible with vasogenic edema, affecting the occipital regions bilaterally in a symmetrical way, also affecting the frontal regions.

medications is recommended (FUGATE JE & RABINSTEIN AA, 2015; LEGRIEL S, et al., 2011; ROTH C & FERBERT A, 2011).

Regarding arterial hypertension, it is known that the gradual reduction in blood pressure levels promotes significant improvement in patients and that the blood pressure goal must be reassessed according to the individual's recovery (RACCHIUSA S, et al., 2019; MCDERMOTT M, et al., 2018; LIMAN TG, et al., 2019). When it comes to malignant hypertension, a reduction in diastolic blood pressure to about 100 to 105 mmHg over an interval of 2 to 6 hours is associated with better outcomes (ANDERSON RC, et al., 2020; FISCHER M & SCHMUTZHARD E, 2017 ; BARTYNSKI WS, 2008).

In most cases, PRES is completely reversible within days to weeks after removing triggers and optimizing blood pressure control. Despite the favorable evolution trend, neurological damage and death were reported as outcomes resulting from PRES (FITTRO K & DIZON R, 2018; LEGRIEL S, et al., 2011).

CONCLUSION

PRES is a neurological syndrome whose typical signs and symptoms include headache, mental confusion, visual symptoms, and seizures, associated with findings consistent with vasogenic edema in the subcortical white matter with a predominance in the posterior cerebral hemispheres seen in neuroimaging exams. Although it occurs more frequently in the context of hypertensive crisis, pre-eclampsia or the use of immunosuppressive medications, PRES manifests itself in many clinical contexts. Treatment of arterial hypertension is the mainstay of treatment and recovery tends to occur within two weeks, but a small proportion of patients remain with residual neurological deficits.

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